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Are catheter intervention and surgery sufficient in patients with tetralogy and comparable lesions?

Konrad Brockmeier

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Nothing to disclose

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PANEL:Sudden Death in Adult Congenital Heart Disease

Chairs: *Juha Matti Happonen, Fabrizio Drago*

* What do epidemiologic studies tell us? *Jane Crosson*
* Noninvasive evaluation methods, ECG, ECHO and MRI. *Canan Ayabakan*
* Risk stratification to avoid unnecessary ICD in patients with structural CHD. *John Papagiannis*
* Are catheter intervention and surgery sufficient in patients with tetralogy and comparable lesions? *Konrad Brockmeier*
* Device implantation: Which device, and how? *Elizabeth Stephenson*

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Tetralogy: 36-years survival of 85%

- Death may occur from re-operation, endocarditis or congestive heart failure
- Sudden cardiac death (SCD) is the leading cause of mortality
- The incidence of VT, death, or both estimated 4.8 events per 100 patient-years

Circulation 2009;119;445
N Engl J Med. 1993; 329:593
Eur JCardiothoracSurg.2007;32:462
J Am Coll Cardiol. 1998;32:245

SCD may be related to the

- underlying anatomical heart defect
- focal scarring
- fibrosis
- surgical suture lines

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Who is at risk for SCD ?

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Risk factors for TOF and VT

- older age at complete repair,
- older age at follow-up,
- history of palliative shunts,
- high-grade ventricular ectopy,
- inducible VT on electrophysiology study,
- abnormal RV hemodynamics
  due to abnormal mechano-electric interaction,
- wide QRS greater than 180 ms

.... no single criterion is highly predictive.

Berul CI, *J Cardiovasc Electrophysiol* 1997

556 TOF patients
11 centers
54.0% female, 36.8+/-12.0 years of age
43.3% with sustained arrhythmia +/or arrhythmia intervention
atrial & ventricular arrhythmias

Khairy et al , Circulation. 2010;122:868-875

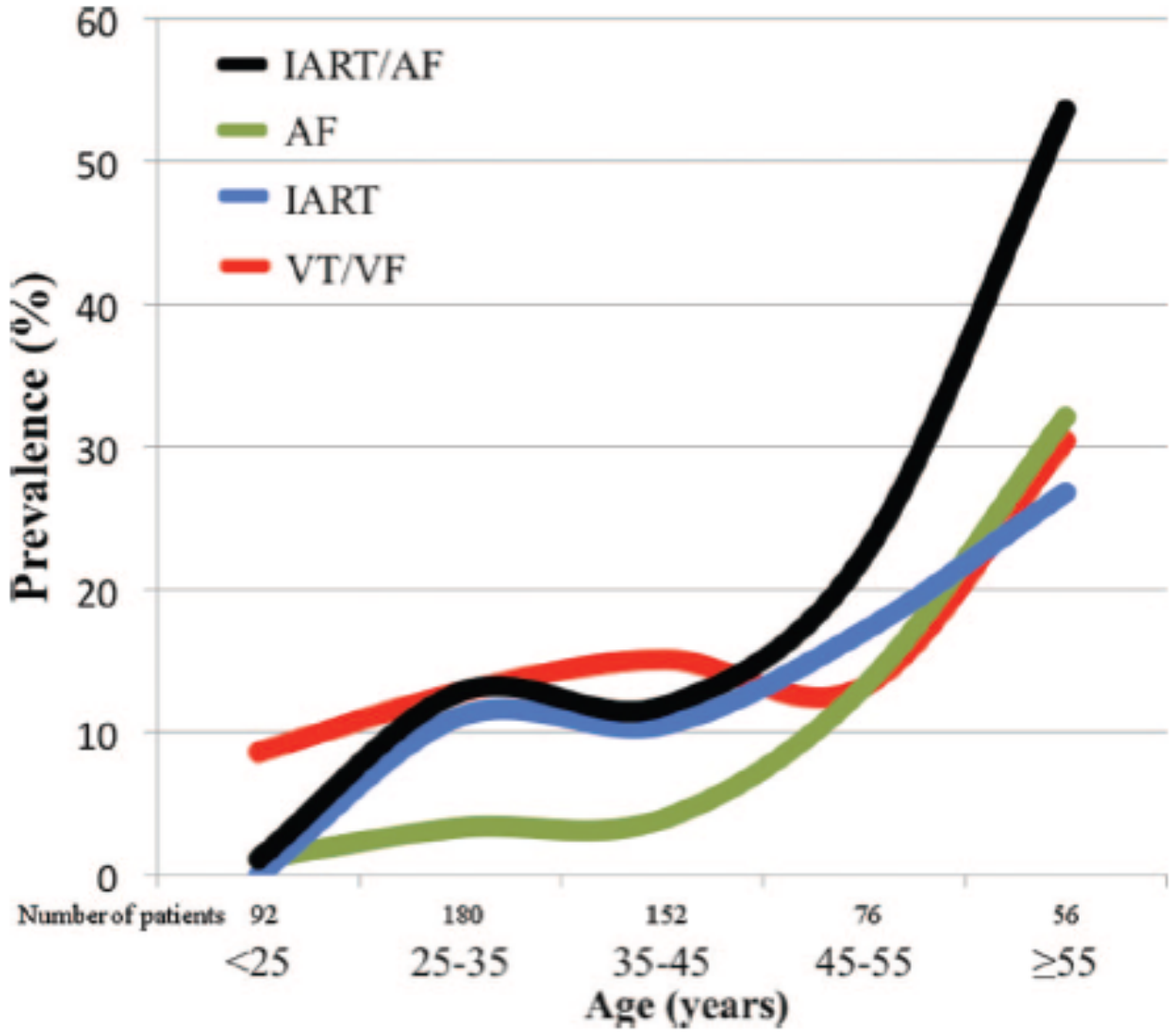

Figure 1. Prevalence of tachyarrhythmias in surgically repaired tetralogy of Fallot according to age. Shown are prevalence rates for IART, AF, IART or AF, and VT or VF by age category. The

Khairy et al , Circulation. 2010;122:868-875

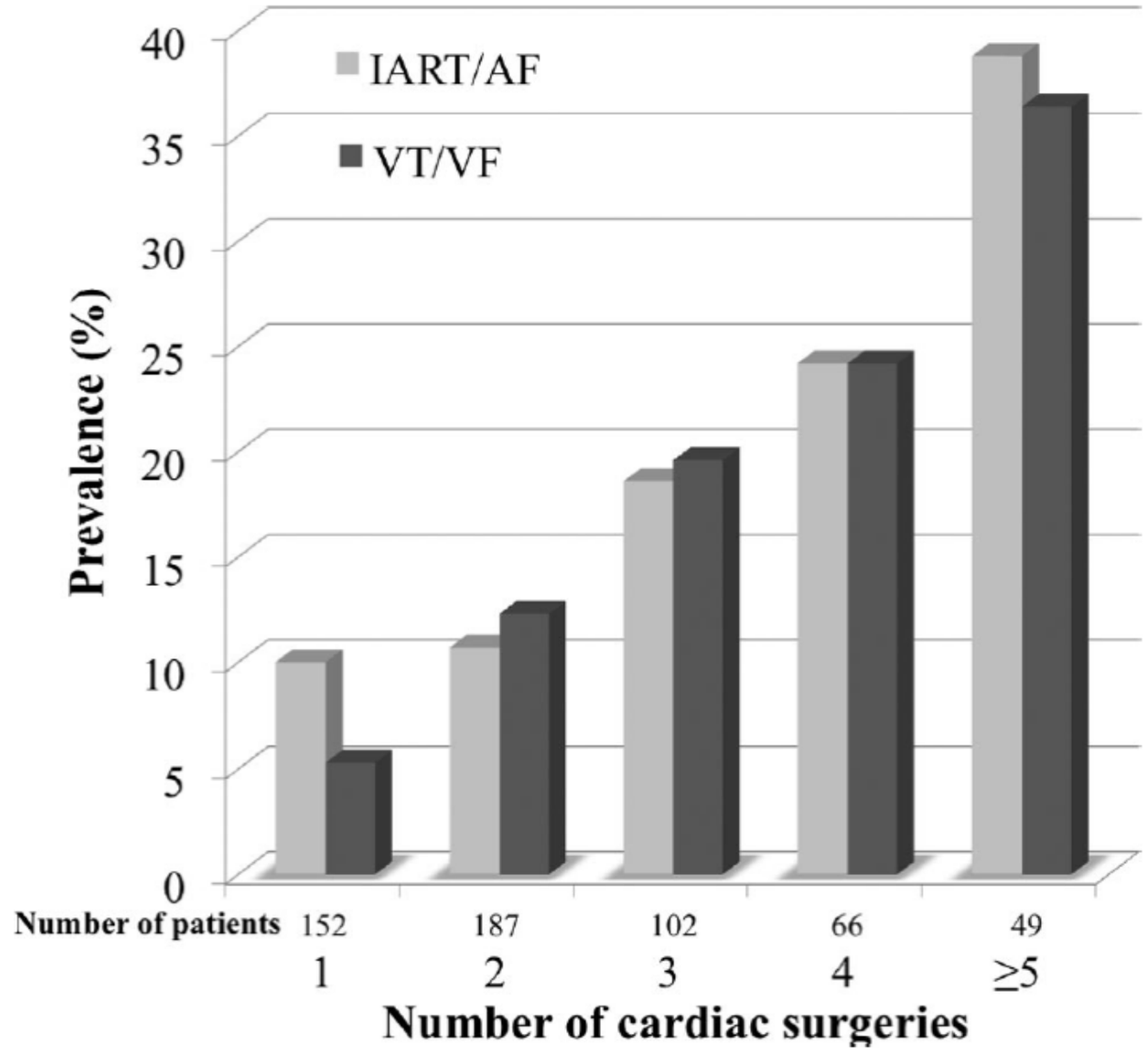

Figure 2. Prevalence of atrial and ventricular arrhythmias according to number of cardiac surgeries. Shown are prevalence rates for atrial arrhythmias, ie, IART or AF, and ventricular arrhythmias, ie, VT or VF, according to number of cardiac surgeries.

Khairy et al , Circulation. 2010;122:868-875

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Details of results

- Median age at corrective surgery 5.0 years (3.0 - 9.0 years)
- Prior palliative shunts 46.6%
- Pulm. transannular patch 80.4%
- RV-to-PA conduits 10.9%
- PV replacements 42.6%

Khairy et al , Circulation. 2010;122:868-875

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Details of results

calculations from multivariate analysis showed highest OR for

- IART .....right atrial enlargement.....OR 6.2
- AF.....left atrial enlargement.....OR 3.2
- VT.....left ventricular dysfunction....OR 3.3

Khairy et al , Circulation. 2010;122:868-875

Impact of surg. pulmonary valve replacement:

Impact of Pulmonary Valve Replacement on Arrhythmia Propensity Late After Repair of Tetralogy of Fallot

J. Therrien, S. C. Siu, L. Harris, A. Dore, K. Niwa, J. Janousek, W. G. Williams, G. Webb and M. A. Gatzoulis

Circulation 2001;103;2489-2494

The relation of clinical arrhythmias and PR is well known ...

Variables	Study group (n=70)
Male sex	33 (47)
Prior palliation	40 (60)
BT shunt	34 (48)
Mean age at repair, y	9.7±8.2
TAP	27 (39)
Mean age at PVR, y	27.8±11.9
Mean time from repair, y	18.1±8.3

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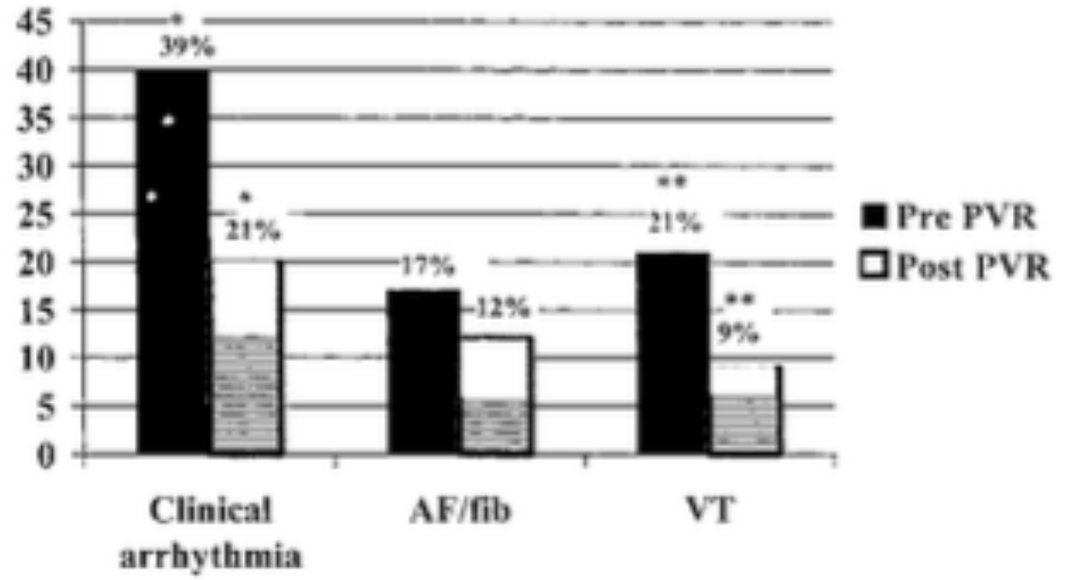

* p= 0.005

** p<0.001

Figure 3. Change in incidence of clinical arrhythmia after PVR. AF/fib indicates atrial flutter or fibrillation; VT, ventricular tachycardia. Dashed area represents de novo arrhythmia after PVR.

Original Article

Ventricular Arrhythmia Risk Stratification in Patients With Tetralogy of Fallot at the Time of Pulmonary Valve Replacement

Anna Sabate Rotes, MD; Heidi M. Connolly, MD; Carole A. Warnes, MD; Naser M. Ammash, MD; Sabrina D. Phillips, MD; Joseph A. Dearani, MD; Hartzell V. Schaff, MD; Harold M. Burkhart, MD; David O. Hodge, MS; Samuel J. Asirvatham, MD; Christopher J. McLeod, MBChB, PhD

Mayo

Background—Most patients with repaired tetralogy of Fallot require pulmonary valve replacement (PVR), but the evaluation for and management of ventricular arrhythmia remain unclear. This study is aimed at clarifying the optimal approach to this potentially life-threatening issue at the time of PVR.

Methods and Results—A retrospective analysis was performed on 205 patients with repaired tetralogy of Fallot undergoing PVR at our institution between 1988 and 2010. Median age was 32.9 (range, 25.6) years. Previous ventricular tachycardia occurred in 16 patients (8%) and 37 (16%) had left ventricular dysfunction, defined as left ventricular ejection fraction <50%. Surgical right ventricular outflow tract cryoablation was performed in 22 patients (10.7%). The primary outcome was a combined event including ventricular tachycardia, out-of-hospital cardiac arrest, appropriate implantable cardioverter defibrillator therapy, and sudden cardiac death. Freedom from the combined event at 5, 10, and 15 years was 95%, 90%, and 79%, respectively. In the first year after PVR, 2 events occurred. Conversely, in the 22 patients who underwent surgical cryoablation, a single event occurred 7 years after PVR. A history of ventricular tachycardia and left ventricular dysfunction was associated with higher risk for the combined event (hazard ratio, 4.7; P=0.004 and hazard ratio, 0.8; P=0.02, respectively).

Conclusions—Patients with repaired tetralogy of Fallot undergoing PVR with history of ventricular tachycardia or left ventricular dysfunction appear to be associated with a higher risk of arrhythmic events after operation. Events in the first year after PVR are rare, and in select high-risk patients, surgical cryoablation does not seem to increase arrhythmic events and may be protective. **(*Circ Arrhythm Electrophysiol*. 2015;8:110-116. DOI: 10.1161/CIRCEP.114.001975.)**

Key Words: arrhythmias, cardiac ■ cryosurgery ■ heart valve prosthesis implantation ■ pulmonary valve ■ tetralogy of Fallot

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n = 205
median age 33 y
VT 8%
LV dysfunction 16%
Freedom from event at 5y = 95%, 10y = 90%, 15y = 79%

—> history of VT and LV dysfunction was associated with higher risk

Rotes AS et al. [***Circ Arrhythm Electrophysiol.*** **2015;8:110-116.**]

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n = 22 had surg. cryoablation

1/22 had VT after 7y

Rotes AS et al. [*Circ Arrhythm Electrophysiol*. 2015;8:110-116.]

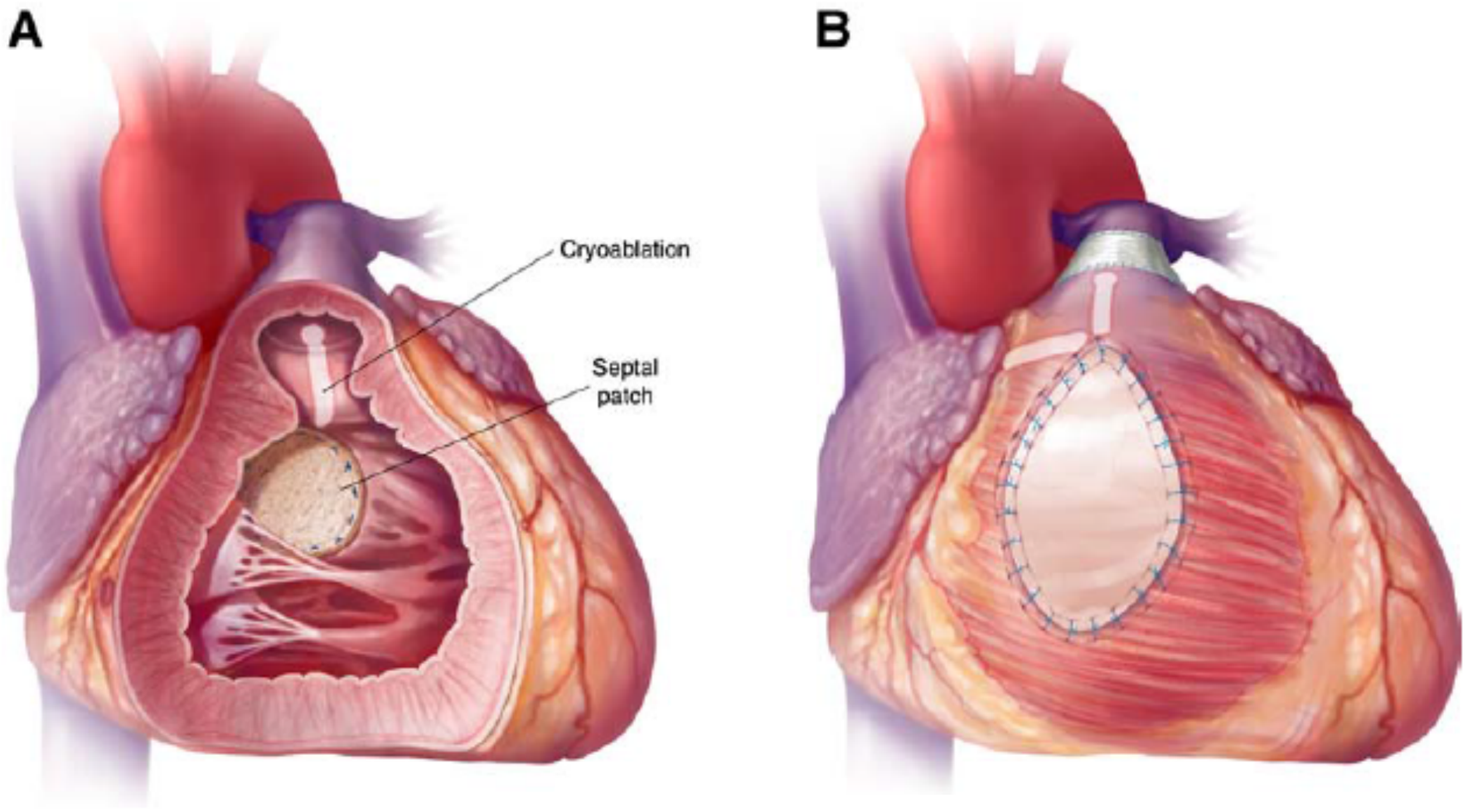

Figure 1. Surgical cryoablation lesions: (**A**) from ventricular septal defect patch, across the pulmonary annulus and up to the pulmonary artery and (**B**) from the right ventriculotomy/patch up to the pulmonary annulus and proximally toward the tricuspid valve annulus level.

Rotes AS et al. [*Circ Arrhythm Electrophysiol*. 2015;8:110-116.]

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Hope

.....a more “aggressive“ approach towards PVR
will improve outcome and reduce arrhythmia burden...?

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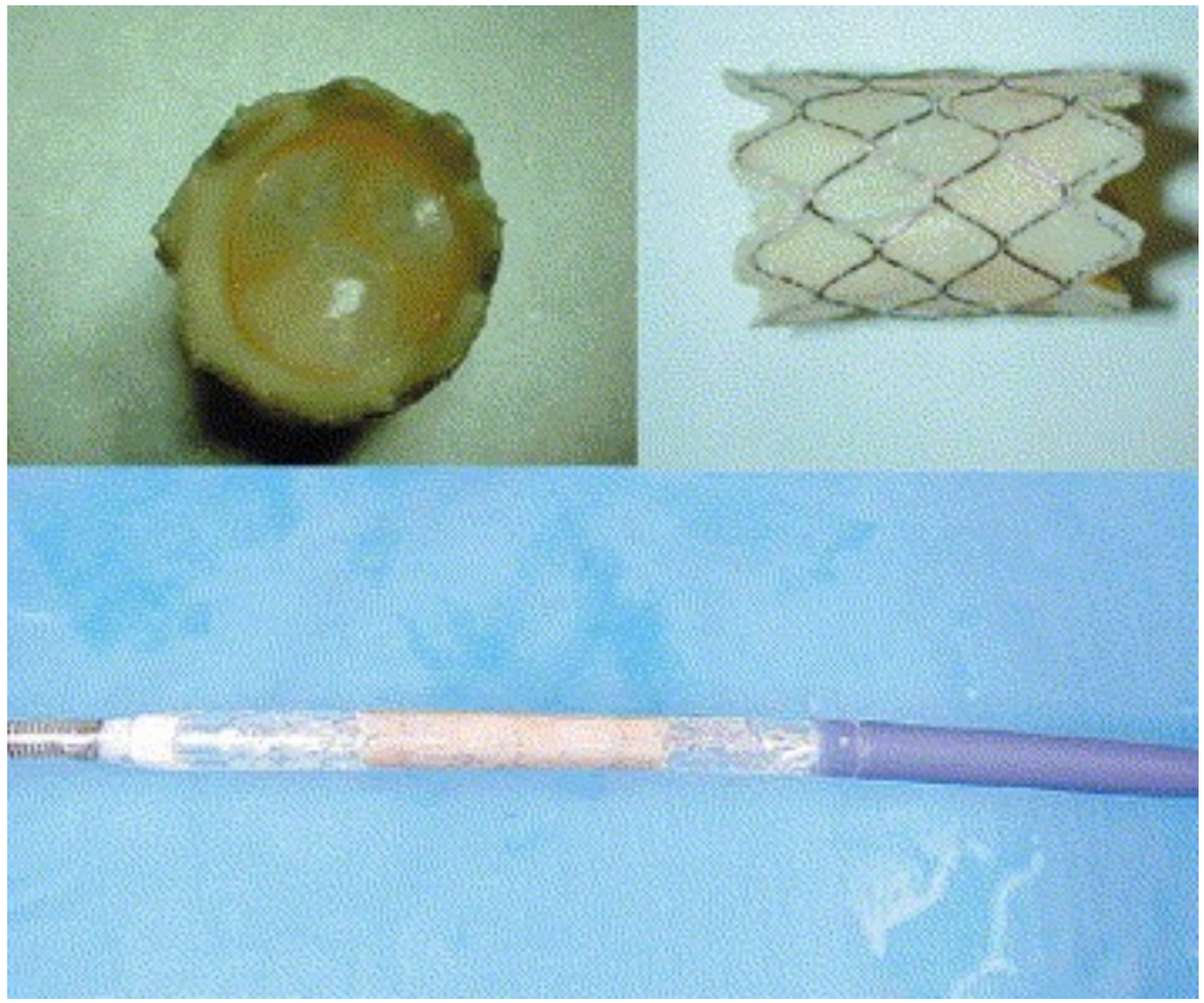
Percutaneous pulmonary valve implantation



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Percutaneous pulmonary valve implantation

n =106
FU 28 months (7-63 months)
pre-implantation
25 patients (24%) had documented arrhythmias:
NSVT …….. n=9, 8%
PVCs …….. n=6, 6%
AF/AFL…… n=10, 9%

Nguyen HH et al. J Am Heart Assoc. 2016

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Percutaneous pulmonary valve implantation

n =106

FU 28 months (7-63 months)

post-implantation

arrhythmias resolved in 4 patients who had NSVT (44%) and
in 5 patients who had PVCs (83%)

Nguyen HH et al. J Am Heart Assoc. 2016

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Percutaneous pulmonary valve implantation

n =106

FU 28 months (7-63 months)

post-implantation FU at 6 month

16 pts had developed new arrhythmias

resolved in 6 pts with NSVT and 7 pts with PVCs

Nguyen HH et al. J Am Heart Assoc. 2016

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Percutaneous pulmonary valve implantation

In line with better hemodynamic results after PPVI

PPVI reduced prevalence of NSVT
most post-implant arrhythmias resolve during FU

Nguyen HH et al. J Am Heart Assoc. 2016

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Animal model from Bordeaux

Optical mapping of rTOF RV-perfused wedges revealed a significant

- prolongation of RV activation time with
- slower conduction velocities and
- regions of conduction slowing

.....well beyond the surgical scar

Benoist D, et al. Heart 2016;0:1–8

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Animal model from Bordeaux

Reduced protein expression of Connexin-43 were identified in rTOF RVs
Remodelling of extracellular matrix-related gene expression
Increase in collagen content correlating with prolonged RV activation time

Benoist D, et al. Heart 2016;0:1–8

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Animal model from Bordeaux

Significant remodelling of RV conduction and repolarisation properties

Remodelling generates a proarrhythmic substrate
likely to facilitate re-entries

—> to contribute to SCD in patients

Benoist D, et al. Heart 2016;0:1–8

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So:

- VT in the FU of TOF surgery should be evaluated both hemodynamically and electrophysiologically
- In many patients cath ablation is possible
- Cryo-ablation during surgery should be considered
- Percutaneous pulmonary valve implantation has to be followed-up on a longer scale

.....some patients will need an ICD implanted

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Are catheter intervention and surgery sufficient in patients with tetralogy and comparable lesions?

... probably not